Extensive congenital abdominal aortic aneurysm and renovascular disease in the neonate

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Primary congenital abdominal aortic aneurysm is an extremely rare entity, with only 15 patients reported in the literature. Options for repair are often limited secondary to branch vessel size and other anatomic limitations. We present a neonate diagnosed with an abdominal aortic aneurysm on prenatal ultrasound. A postpartum computed tomography angiogram revealed an extensive type IV thoracoabdominal aortic aneurysm extending to the aortic bifurcation and resulting in bilateral renal artery stenosis. The unique features of this patient and challenges in management are discussed. (J Vasc Surg 2012;55:1762-5.)

Abdominal aortic aneurysm (AAA) in neonates is rare and occurs most commonly secondary to other causes such as connective tissue disease, infection, vasculitis, and umbilical artery catheterization.1 Primary congenital AAA is very uncommon, with only 15 such aneurysms diagnosed in infancy having been reported thus far.2-16 The paucity of reported cases precludes a standardized approach to management of these aneurysms. Although repair has been successful in several patients,2,8,10,12,15 aneurysm extent and anatomy often prohibit repair in neonates. In addition, concomitant renovascular hypertension can further complicate management in these patients.5,7,11 Here we describe a neonate diagnosed prenatally with what appears to be the most extensive AAA yet reported. The obstacles presented in association with nonoperative management in the setting of renovascular disease are discussed.

CASE REPORT

A girl, weighing 2472 grams, was born by elective Cesarean section at 34-4/7 weeks’ gestation. She was twin B in a dichorionic-diamniotic twin pregnancy to a 24-year-old G4 P0030 mother. An ultrasound examination at 32 weeks’ gestation had revealed an AAA with possible dissection flap. No abnormal findings were noted in the sibling. Apgar scores were 7 at 1 minute and 8 at 5 minutes. She required blow-by oxygen briefly after birth and was transferred to a tertiary children’s hospital for further evaluation of the AAA.

A computed tomography angiogram revealed a fusiform aneurysm measuring 2.3 cm in maximum diameter and extending from just above the diaphragm to the aortic bifurcation (Fig 1). There were luminal folds distally but no true dissection. All visceral vessels originated from the aneurysm with evidence of ostial stenosis. The renal arteries were very small, and no obvious connection with the aortic lumen was seen on computed tomography angiogram. The right renal artery appeared to be fed primarily through phrenic artery collaterals, and both kidneys showed minimal contrast enhancement (Fig 2). Abdominal ultrasound imaging revealed patent renal and mesenteric vessels but with flow patterns consistent with ostial effacement. A prenatal and two postnatal echocardiograms demonstrated mild right ventricular hypertrophy and no evidence of aortic dissection.

After multidisciplinary discussion involving neonatology, surgery, genetics, and nephrology, it was determined that repair would require complete replacement of the abdominal aorta and reimplantation of all branch vessels. Given the patient’s size relative to the aneurysm and the congenital stenosis of the branch vessels, we elected to manage nonoperatively with aggressive blood pressure control and defer surgical intervention.

Upon presentation to our hospital she had a mean arterial pressure (MAP) of \( \approx 90 \) mm Hg. She was initially started on an esmolol drip to control her hypertension. The decision was made to avoid angiotensin-converting enzyme inhibitor or angiotensin receptor blocker therapy in the setting of severely impaired renal perfusion on imaging. The patient was transitioned to isradipine and weaned off esmolol once MAPs were maintaining near 70 mm Hg.

During her admission, a discussion was held with the parents regarding the complexity of the aneurysm and the dire consequences of rupture. She was discharged on isradipine for blood pressure control (goal MAP <70 mm Hg) to reduce the risk of aneurysm expansion and rupture. Transforming growth factor-\( \beta \) receptor 1 and 2 DNA sequencing revealed no mutations, thus excluding Loeys-Dietz syndrome as a cause of the patient’s aneurysm.

The plan was to closely monitor blood pressure and renal perfusion as an outpatient with a plan for reimaging and possible repair at age 6 months, when the patient’s growth might make her vascular anatomy more amenable to operative intervention. Before discharge, the family decided to make the patient do not resuscitate. She was noted to have good blood pressure control at home and at her nephrology follow-up appointment. At age 4 weeks, a few hours after a follow-up appointment with her pediatrician, the patient presented to another emergency department with a firmly distended abdomen and unrecoverable...
blood pressure. She ultimately died, with the clinical diagnosis of a ruptured aneurysm.

DISCUSSION

Since the first case report describing a ruptured AAA in infancy,9 only 14 other patients have been reported, most of which were infra ovarian.1,2,6,8,10,12,16 This is only the second report of a congenital type IV thoracoabdominal aneurysm. Buddingh et al3 described a neonate with several saccular aneurysms combining to form a large thoracoabdominal aneurysm, but to our knowledge, the present patient represents the most extensive continuous congenital AAA yet reported and the first with severe visceral and renal artery stenosis producing renovascular hypertension.

Although it is generally accepted that early elective repair is preferred to prevent rupture, no standard operative approach has been established. Multiple repair methods have been reported, including aneurysmorrhaphy,15 repair with native vessels,12 cryopreserved allograft,2,14 and various synthetic grafts.8,10,13 Timing of these repairs range from the neonatal period up to age 15 months, but all were undertaken in patients whose AAAs were confined to the infrarenal aorta. No repair was attempted in several patients due to aneurysm extent and prohibitively small vessel size3,5,7,11,16 as well as hemodynamic instability from concomitant medical issues.7,11 Two patients presented with acute rupture and did not survive operative intervention6,9 (Table). Our patient was hypertensive, and the extent of the aneurysm and small size of the splanchnic vessels made repair impractical. This further highlights that although prenatal diagnosis of AAAs is helpful in identifying patients who may benefit from early operative intervention,4,8,11,16 vascular anatomy may preclude such intervention.

Nonoperative management must aim to minimize the risk of aneurysm expansion and rupture. Unfortunately, the discussion of nonoperative approaches in these patients in the literature is even sparser than that addressing operative management. Therapeutic decisions are largely extrapolated from the adult literature. The cornerstone of long-term management in adults is blood pressure control. The logic behind this approach is that aortic wall stress is higher in AAAs that progress to rupture, and that antihypertensive therapy decreases wall stress.17,18
Complicating the present case was that the patient had severely impaired renal perfusion contributing to her elevated MAPs. Renovascular hypertension has been reported in two other patients with congenital AAA, both of whom died of concomitant conditions within the first weeks of life. This patient required careful decision making regarding antihypertensive therapy to balance her risk of aneurysm expansion with her renovascular disease. Given the risk of progressive azotemia with angiotensin-converting enzyme inhibitor use in the setting of renal dysfunction, the patient was started on esmolol and transitioned to isradipine. Calcium channel blockers were used in one other patient with congenital AAA and showed at least some efficacy in the treatment of AAA in animal models, although the benefits of specific drugs in pediatric AAA management has not been investigated.

**CONCLUSIONS**

This patient presented with the most extensive continuous congenital AAA yet reported. The case highlights a number of important factors in the management of these aneurysms in neonates, especially with regard to anatomic considerations and hypertension management in patients with concomitant renovascular disease. It further stresses the importance of an individualized approach in treating neonates with this rare entity.

**REFERENCES**


